SOME ATYPICAL FORMS OF TABES AND PARESIS CONSIDERED IN THE LIGHT OF SERODIAGNOSIS¹

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The corroborative value of serodiagnostic methods in the study of irregular types of parasyphilitic disease is beyond question. Cerebrospinal syphilis and parasyphilis are basically different states. Mott makes this distinction very clear. When Crocq reports the cure of incipient tabes and early paresis it is nervous syphilis, not tabes or paresis, that is cured. Serodiagnosis should have prevented the confusion of these diseases, although there are very exceptional cases in which the differentiation is difficult to make. Every case of cerebrospinal syphilis, tabes or paresis should have a Wassermann made of both blood and spinal fluid, also a lymphocyte count and globulin test (the four clinical reactions of Nonne), since it is only in this way that the presence of syphilis may not be overlooked, and the different form of syphilogenous disease differentiated.

The following cases will best illustrate this:

CASE I. Some five years ago E. consulted me for periodic attacks of nausea, vomiting, and pain, accompanied by great prostration, which would after a time cease and apparent restoration to health take place. Examination of both stomach and nervous system was negative; there was however a marked lymphocytosis. Evidently this was a case of tabes of which the gastric crises were the one and only symptom. This is in accord with Nonne's dictum that in parasyphilitic disease there is always more or less pleocytosis.

A propos of this case is one reported by Plaut of a woman alcoholic, 44 years of age, in whom there was inequality of the pupils with left-side immobility; patellar reflexes were normal. There were no other symptoms. There was no lymphocytosis but the spinal fluid gave a positive Wassermann. One year later a classical tabes had developed.

CASE II. B., aged 50, Canadian, widower, merchant, two children living and well; wife had one miscarriage. Contracted

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syphilis 12 years ago; six years later he greatly exhausted himself by rowing and a short time afterwards he felt tingling in the left little finger which spread to all the fingers. Occasionally there were shooting pains. One year later a similar condition developed in the right hand. For the past year there has been difficulty in writing and in buttoning his clothes. For four years there has been a loss of sexual power. No bladder disturbance. Shooting pains are sometimes felt in legs; no Argyll-Robertson pupil. Traumatic cataract of the left eye. Knee jerks normal, Achilles jerks absent, slight Rombergism and marked ataxia of both hands. Biernacki's sign present. There was a band of hyperesthesia in the distribution of the fifth cervical nerve of both arms. Pressure of the spinal fluid increased; Nonne-Apelt and Noguchi positive; very marked lymphocytosis; blood serum negative; spinal fluid gave a positive Wassermann. This reaction is not in accord with that usually occurring in tabes; however Plaut states "it would seem that in tabes as opposed to my findings in paresis, cases do occur which present a negative behavior of the serum."

CASE III. Mrs. W., aged 43, Swede, housewife, one miscarriage at third month, one child living and well. Has had migraine for the past fifteen years. Was operated upon for gall stones in 1910. Headaches have been very severe for the past two years; pain would last twelve hours, then nausea and vomiting would occur for from two to four days. At first these attacks were from three to four months apart, but now they come on every few weeks. During a seizure the patient is unable to void urine and has to be catheterized; she is much depressed at these times. With each migrainous attack there is associated a typical Raynaud syndrome which comes on with the vomiting. No Argyll-Robertson pupil, although the reaction to light is somewhat sluggish. Beginning atrophy in the left optic nerve, right normal; marked Romberg; no ataxia of upper extremities. Last year there were shooting pains in the legs. Deep and superficial reflexes normal; no sensory disturbances; blood pressure 100 mm. Hg., hemoglobin 91 per cent., red blood cells 4,600,000; albumin in urine but no casts. Increased pressure of spinal fluid; no lymphocytosis; Nonne-Apelt and Noguchi positive; blood serum gave a positive Wassermann, spinal fluid doubtful. The occurrence of the Raynaud syndrome is very unusual and I have never before seen it in tabes. Crocq reports a case seen by Babinski in an heredity syphilitic. Gaucher, Claude and Croissant refer to its occurrence in a young man some months after the appearance of the initial sore, and which was cured by antisyphilitic treatment.

CASE IV. H., 47 years, American, physician, excessive smoker; had typhoid in 1887; wife had one miscarriage, two healthy children. Contracted lues in 1898. Ten months ago first

felt soreness in the soles of both feet, which was most noticeable at night. Shortly after numbness developed in the toes and in the feet. There was also aching in the calves of both legs. Similar sensory disturbances then manifested themselves in both arms. No bladder trouble or loss of sexual power. He complained of vague pains in the masseter muscles. No Argyll-Robertson pupil, no ataxia, no Romberg. Knee jerks normal, Achilles jerks normal, sensation normal. Increased pressure of spinal fluid. Nonne-Apelt and Noguchi tests negative; no lymphocytosis. Wassermann in the cerebrospinal fluid positive, in blood serum negative.

The three characteristic tabetic symptoms are Argyll-Robertson pupil, loss of knee jerks and lightning pains. Strümpell states if two of these, especially if one be the Argyll-Robertson pupil, be present the case is very probably tabes. In my four cases the Argyll-Robertson pupil was absent in all, the knee jerks were normal in all, the lightning pains were present in two cases. In case one there was one symptom only, gastric disturbance; in case two, momentary pains, slight Rombergism, Biernacki's sign, ataxia of the hands and hyperesthesia in the distribution of the fifth cervical nerve in both arms. In case three there were sluggish pupils, beginning atrophy of the optic nerve, shooting pains in legs and Rombergism. In only two of these patients was a history of syphilis elicited. The absence of the knee jerk is one of the most constant symptoms of tabes (95 per cent. of the cases, Erb). We have all seen it present in the beginning of the disease and then disappear sometimes following an attack of lightning pains; or one may be present and the other lost. This Mott regards as suggestive of tabo-paralysis. In rare cases it may be retained throughout the disease (Erb). The Achilles jerk was normal in three cases, absent in one. While this is usually lost before the knee jerk, occasionally the converse obtains. Erb regards its absence, especially its unilateral absence, just as important a symptom as that of the patellar reflex. In all of these patients the Argyll-Robertson pupil was wanting.

Does this phenomenon bear a definite relation to the tabetic syndrome? Erb says it is the earliest and most frequent symptom of tabes; Williamson that it is a very early sign and is found in two-thirds of the cases; Oppenheim that it ranks with the knee jerk, that frequently it is the first definite symptom, and that he

has observed cases where for ten or fifteen years it was the only objective sign; and Fisher, that he would hesitate to make a diagnosis of tabes or paresis if it were absent. On the other hand Babinski states that the Argyll-Robertson pupil is simply an indication that the central nervous system has been touched with syphilis. DeMassery believes that this symptom is neither pathognomonic nor constant in tabes. Siemerling found it present in only one per cent. of his 1639 cases of nervous syphilis, and Clark observed it in five patients out of sixty-nine suffering from this disease. According to Henderson it is present in 70 per cent. of all cases of tabes and paresis. Babinski has called attention to its association with lymphocytosis. The presence of the Argyll-Robertson pupil then has absolutely no differential diagnostic value, it simply indicates a prior luetic infection, and a majority of the patients so affected manifest, unfortunately, a distinct predilection to neuronc degeneration. In case three there was beginning atrophy of left optic nerve with lightning pains in the legs. In rare cases, as in this instance, optic atrophy may occur with normal knee jerks. It is an early symptom in 2 per cent. of the cases (Williamson). It is observed in from 10 to 15 per cent. of tabetics (Erb, Mott) and is frequently followed by tabo-paralysis. It may be the only symptom or it may be preceded by lightning pains, or it may be associated with these and loss of knee jerk. Generally the Argyll-Robertson pupil is present.

A study of the cytologic, biologic and globulin reactions in these 4 cases, and a comparison of the findings with the usual reactions occurring in tabes dorsalis is not without interest and is of much practical value. In case I there was only a cytologic examination, since at the time the brilliant researches of Wassermann and his co-workers were yet in the making. In case two and three and four lymphocytosis was positive once, negative twice; the globulin reaction was positive twice, negative once. A positive globulin reaction is more constant than the cellular increase in tabes and paresis, and it is entirely missing in lues when the latter is uncomplicated by lesions of the nervous system (Holzmann). The globulin reaction occurred with greater frequency in these cases. Both of these reactions are however constant factors in tabes.

The biologic reaction of the blood serum was negative twice, positive once; the cerebrospinal fluid, positive twice and doubtful

once. These findings are not only wanting in uniformity but also fail to accord with the types of reaction believed to be characteristic of tabes, viz., a positive reaction of blood serum and cerebrospinal fluid, with lymphocytosis and a positive globulin reaction, although according to Plaut the reacting substances are not as constantly present in the blood serum and cerebrospinal fluid as in paresis. On the other hand, Kaliski reports the blood serum to be positive in from 60 to 70 per cent., and the spinal fluid in from 10 to 20 per cent. of the cases; while Holzmänn states that the blood serum is positive in 70 per cent., the globulin reaction in 95 per cent., and lymphocytosis in 90 per cent. of the cases. The spinal fluid (original method) is positive in from 5 to 10 per cent.; larger quantities of fluid in nearly 100 per cent. of the cases. Sometimes the biologic reaction in tabes, as in case two (blood serum negative, spinal fluid positive), corresponds to an atypical reaction which I have observed in cerebral lues. There seems to be a consensus of opinion among neurologists that lymphocytosis is an early and constant symptom of tabes (Strümpell, Erb, Oppenheim, Mott, Nonne). DeMassery says that he has known certain rare cases of tabes, at least in certain periods of their course, without lymphocytosis. The case of atypical paresis which I now report is sufficient for the purpose of this paper.

In this patient all of the four reactions which Nonne believes almost invariably indicate paresis were present. The clinical syndrome was in no manner distinctive of paresis but was rather suggestive of manic-depressive insanity; "that vile phrase" as one of the distinguished members of this society calls it.

G., male, American, aged 48, married, no nervous or psychopathic heredity; no previous mental breakdown. Became luetic at 28. He was an excessive smoker and drinker. Wife had one miscarriage; one child died of hydrocephalus, one living. Two months before consulting me he began to suffer from insomnia and periods of excitement and confusion during the night; rational in the daytime. He thought dead bodies were around him, heard people moving coffins in the basement, etc., speech thick, very irritable and unmanageable, confused and disoriented. There was no elation nor grandiose ideas. The neurological and physical examinations were negative. Hemoglobin 84 per cent. red blood cells 4,864,000, white cells 8,975; blood pressure 146 m.m.Hg. Urine normal, a positive Wassermann of both serum and spinal fluid; mild lymphocytosis; Nonne-Apel't test positive. He grew worse, became bedridden and apparently was rapidly

deteriorating; he was given mercurial inunctions with iodide and increasing doses of tuberculin after the method of von Wagner of the Vienna Psychiatric Clinic. After eight weeks, treatment was stopped and the patient was removed to a state hospital. Shortly after this he began to improve and at this writing is presumably well. The asylum superintendent said it was a manic-depressive case and not paresis. Some months after leaving the asylum I received a letter from the patient; it was clear, and free from any evidence of mental impairment, but the writing was very suggestive of paresis. Holzmann states that in a pathologic spinal fluid there is increased pressure, a positive globulin reaction and increased cell content; and if these three symptoms are present either in combination or singly we have to do with an organic disease of the nervous system, specific or non-specific. In the three tabetics and the one case of paresis there was increased pressure. Holzmann also says that aside from leprosy the spinal fluid reacts positively only in luetic cases. This reaction was doubtful in only one of my cases and even this case was probably positive.

CONCLUSIONS

I. These cases are certainly anomalous in their findings and emphasize the necessity of testing for the four reactions of Nonne in every instance, for without this no examination can be regarded as final.

II. While probably in typical tabes the serologic response in a majority of cases is in accord with Plaut's findings (blood serum and spinal fluid positive), yet that such is not the fact in incipient cases is clearly shown by these irregular types, and Kaplan has demonstrated that even in uncomplicated cases "no hard and fast lines can be laid down" since in 2 out of 167 patients he obtained a negative result in every one of the "four-phase reactions."

III. The presence of Nonne's four reactions leads me to regard the last case as one of paresis notwithstanding the atypical clinical syndrome and the apparent recovery. The weight of evidence is certainly in favor of paresis.

IV. The cytologic, biologic and chemical findings are not, as Karpas well says, to be considered as the "ultimate court of appeals" for settling disputed diagnoses in doubtful or atypical cases; rather they should be used in conjunction with the clinical syndrome and regarded as corroborative only. Consideration of both these factors will enable one to arrive at a diagnosis in the great majority of instances.